



TITLE:

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COEXISTENCE OF RENAL CELL CARCINOMA AND RENAL ANGIOMYOLIPOMA DEVELOPING IN A KIDNEY: A CASE REPORT

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Coexistence of renal cell carcinoma and angiomyolipoma in the same kidney is rare. A 54-year-old woman without tuberous sclerosis was admitted for further examination of incidental renal masses on ultrasonography. Computerized tomography revealed a 17-mm high density mass in the mediolateral portion of the right kidney and a 5-mm low density mass near the right lower pole. Because the former mass showed a typical tumor pattern on selective renal angiography and the latter mass was strongly hyperechoic on ultrasonography, a clinical diagnosis of renal cell carcinoma and angiomyolipoma was made. A right radical nephrectomy confirmed the preoperative diagnosis. She has been followed for 61 months with no recurrence.

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Key words: Renal cell carcinoma, Angiomyolipoma, Coexistence in a kidney

INTRODUCTION

Renal angiomyolipomas (AMLs) are considered benign mesenchymal tumors, arising from embryonal cells in the kidney. They are found in 40 to 80% of patients with tuberous sclerosis complex¹⁾. Renal cell carcinomas (RCCs) are major malignant tumors accounting for 89% of all renal tumors²⁾. The association of these two entities in the same kidney is uncommon, and it is difficult to distinguish AMLs from RCCs without sophisticated imaging techniques³⁾.

Here, we report a case of renal cell carcinoma and an angiomyolipoma occurring as two separate masses within the same kidney without tuberous sclerosis. The patient was diagnosed preoperatively utilizing computerized tomography (CT), magnetic resonance imaging (MRI), and angiography (AG). To our knowledge, 30 cases of simultaneous renal cell carcinoma and renal angiomyolipoma in the same kidney with or without phacomatosis have been reported previously.

CASE REPORT

A 54-year-old woman was hospitalized for further examination because a hyper-echoic mass in the mediolateral portion (Fig. 1A) and a second mass which was much more hyper-echoic than the former near the lower pole (Fig. 1B) were noted on an ultrasonogram of the right kidney in December 1989. She had no personal or family history of tuberous sclerosis. The abdomen was soft and no masses were palpable. No edema or lymphadenopathy was noted. Neurological examination disclosed no abnormalities. Tuberous sclerosis was not detected.

Laboratory data, including complete blood count, blood chemistry studies and urinalysis were within the normal range except for a slightly elevated erythrocyte sedimentation rate of 25 mm per hour (normal 2 to 12). Excretory urogram revealed a mass in the mediolateral portion of the right kidney without distortion of the collecting system and the left upper urinary tract appeared normal. CT revealed a high density mass of 17 mm in diameter in the right mediolateral portion with mosaic enhancement by contrast medium and a homogeneous low density area of 5 mm in diameter near the right lower pole (Fig. 2). These two tumors were also recognized on MRI (Fig. 3). Transfemoral selective renal AG showed a typical tumor staining pattern in the mediolateral portion of the right kidney consistent with renal cell carcinoma (Fig. 4). Abdominal ultrasonography failed to provide further information. From these examinations, a clinical diagnosis of coexistent RCC and AML in the same

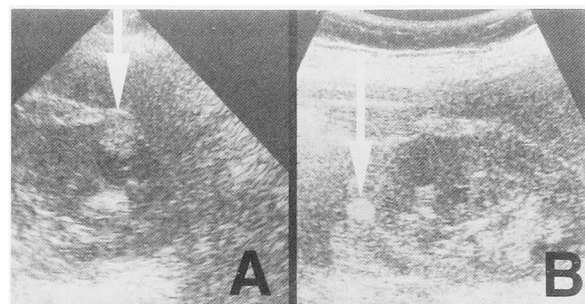


Fig. 1. US findings in the right kidney: A, a hyper-echoic mass in the mediolateral portion (arrow). B, a second mass which was much more hyper-echoic than the former near the right lower pole (arrow).

kidney was made.

In March 1990, a right radical nephrectomy was performed by the transperitoneal approach through a

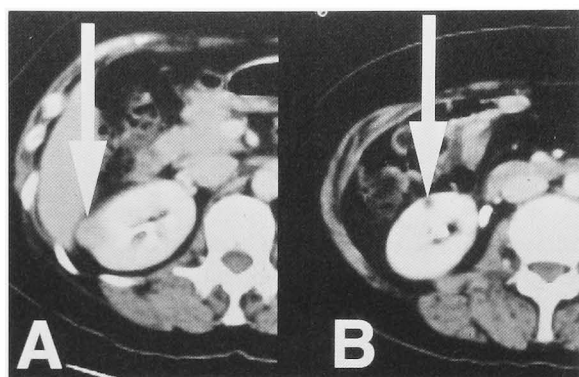


Fig. 2. CT findings in the right kidney: A, high density mass in the right mediolateral portion (arrow). Note this tumor was enhanced heterogeneously by contrast medium. B, low density area near the right lower pole (arrow).

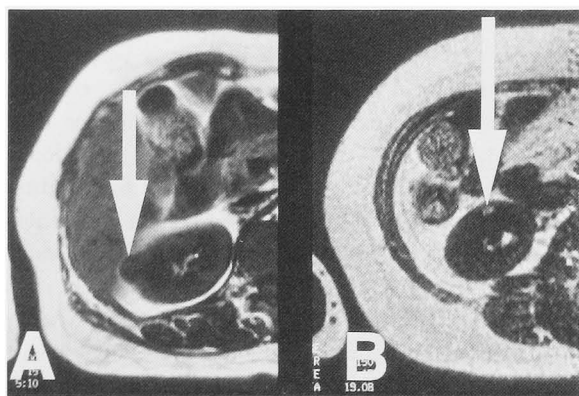


Fig. 3. A, Low intensity area (arrow) in the mediolateral portion of right kidney was displayed on T1-weighted MRI. B, High intensity tumor (arrow) near right lower pole was also displayed on T1-weighted MRI.

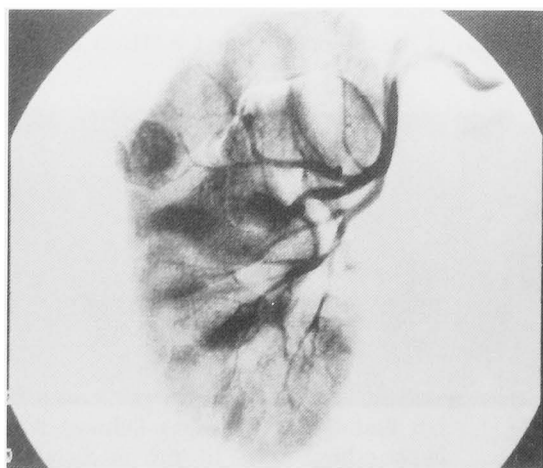


Fig. 4. Selective renal AG demonstrated typical tumor staining pattern in the mediolateral portion of the right kidney.

midline incision. Macroscopically, there were two separated and well circumscribed tumors within the kidney. The mass of the mediolateral portion of the right kidney was variegated yellow to light brown, while the mass near the lower pole was a homogeneous gray-pink. Pathological examination revealed renal cell carcinoma, alveolar type, common type, clear cell subtype, G1 (mass in the mediolateral portion of the right kidney) and renal angiomyolipoma (mass near the lower pole of the right kidney) (Fig. 5). The patient convalesced uneventfully and has been followed for 61 months without any local recurrence or distant metastasis.

DISCUSSION

RCCs comprise approximately 1% of all malignant tumors excluding skin malignancies. RCC most commonly develops in the fifth to seventh decade of age²⁾. AMLs are uncommon benign tumors that arise from embryonal cells in the kidney and are composed of thick-walled blood vessels, smooth muscle and mature adipose tissue⁵⁾. They are known to affect 40 to 80% of patients with tuberous sclerosis, although such tumors can be found in a significant number of otherwise healthy persons¹⁾.

To date, 30 cases of angiomyolipoma accompanied by renal cell carcinoma have been reported as shown in Table 1^{3,4,6-29)}. Seventeen of those cases involved tuberous sclerosis (TS), and these patients were between 11 and 57 years old with a mean age of 31 years old. Seventy-one percent of those with TS were female. However, 14 patients, including ours, did not show TS, and they were between 39 and 74 years old, with a mean age of 53 years old. Sixty-four percent of those without TS were female. Until

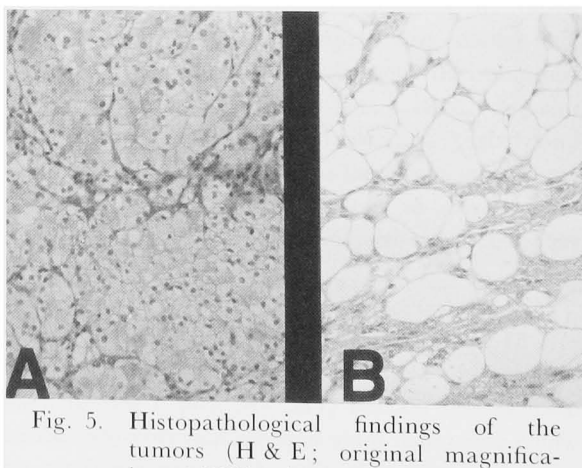


Fig. 5. Histopathological findings of the tumors (H & E; original magnification, $\times 200$): A, renal cell carcinoma, alveolar type, common type, clear cell subtype, G1 (mass in the mediolateral portion of the right kidney). B, mixture of adipose tissue, blood vessels, and smooth muscle in the angiomyolipoma (mass near the lower pole of the right kidney).

Table 1. Documented cases of renal cell carcinoma and angiomyolipoma in the same kidney

Reference	Age/Sex	TS	AML	RCC	AML & RCC in the same mass?	Treatment	Outcome (M)
Jochimsen et al. (1969)	31/F	(+)	Bil	Bil	?	Bil. Nx	48 NED
Kavaney & Fielding (1975)	48/F	(-)	Lt	Lt	No	Lt. Nx	12 NED
Barbour & Casali (1978)	28/M	(+)	Bil	Bil	No	Bil. Nx	10 NED
Gutierrez et al. (1979)	24/F	(+)	Bil	Lt	No	Lt. Nx	13 CD
Lynne et al. (1979)	27/F	(+)	Bil	Bil	Yes	Bil. Nx	42 NED
Mazeman et al. (1980)	50/M	(-)	Lt	Lt	No	Lt. Nx	48 NED
Mazeman et al. (1980)	11/M	(+)	Rt	Rt	?	Rt. Nx	48 NED
Mazeman et al. (1980)	13/F	(+)	Lt	Lt	No	Lt. Nx	36 NED
Schujman et al. (1981)	62/F	(-)	Rt	Rt	Yes	Rt. NX	?
Garini et al. (1981)	22/F	(+)	Bil	Lt	No	Bil. Nx	?
Takeyama et al. (1982)	42/M	(-)	Lt	Lt	No	Lt. Nx	?
Ishii et al. (1984)	48/F	(-)	Rt	Rt	No	Rt. Nx	36 NED
Silpananta et al. (1984)	74/F	(-)	Bil	Lt	No	Lt. Nx, Rt. pNx	?
Graves & Barnes (1986)	29/M	(+)	Rt	Rt	No	Rt. Nx	0.5 DOD
Ahuja et al. (1986)	34/F	(+)	Bil	Bil	No	Lt. Rad, Rt. Emb	12 CD
Ueda et al. (1987)	48/M	(-)	Lt	Lt	No	Lt. Enucl	?
Huang et al. (1988)	39/M	(-)	Bil	Rt	Yes	Rt. Enucl	8 NED
Morita et al. (1988)	49/M	(-)	Lt	Lt	No	Lt. Nx	40 CD
Blute et al. (1988)	62/F	(-)	Rt	Rt	No	Rt. Nx	36 CD
Blute et al. (1988)	73/F	(-)	Lt	Lt	?	Lt. Nx	60 NED
Blute et al. (1988)	38/F	(+)	Bil	Lt	?	Lt. Nx	12 NED
Satoh et al. (1988)	49/F	(+)	Rt	Rt	Yes	Rt. Nx	106 NED
Taylor et al. (1989)	15/M	(+)	Bil	Bil	?	Bil. Nx	24 NED
Wei et al. (1990)	39/F	(-)	Lt	Lt	No	Lt. Nx	?
Yamanaka et al. (1990)	25/F	(+)	Bil	Rt	No	Lt. pNx, Rt. Nx	?
Ohigashi et al. (1991)	37/F	(+)	Bil	Lt	No	Lt. Enucl & pNx	6 NED
Hardman et al. (1993)	50/F	(+)	Rt	Rt	?	Rt. Nx	11 DmAML
Canzonieri et al. (1993)	54/F	(-)	Rt	Rt	No	Rt. Nx	15 NED
Conrad et al. (1994)	57/F	(+)*	Lt	Lt	No	Lt. Nx	12 NED
Nakamura et al. (1994)	40/M	(+)	Bil	Lt	No	Lt. Nx	11 DTS
Present case	54/F	(-)	Rt	Rt	No	Rt. Nx	61 NED

Bil, bilateral; Lt, left; Rt, right; Nx, nephrectomy; pNx, partial nephrectomy; Emb, embolization; Rad, radiation; Enucl, enucleation; NED, no evidence of disease; CD, cancer death; DOD, died of other disease; ?, not documented in the literature; DmAML, died of malignant AML; DTS, died of tuberous sclerosis.

*Conrad et al. classified lymphangiomyomatosis as a forme fruste of tuberous sclerosis.

recently, definitive preoperative diagnosis of renal AMLs was rarely established because of the difficulties in ruling out renal malignancies. Contemporary advances in US and CT have provided a new dimension in evaluating of renal tumors. When fat is demonstrated by these newer radiodiagnostic procedures, we are able to diagnose AML. Whether the RCC and AML occur in the same mass or are separated is important from the perspective of preoperative diagnosis. Urologists and radiologists before Takeyama and associates⁶⁾ could not make a correct preoperative diagnosis when RCC and AML coexisted in the same kidney. To date, however, no one has yet correctly diagnosed coexisting RCC-AML in the same mass preoperatively. Lynne et al.⁷⁾ and Huang et al.⁸⁾ misdiagnosed coexisting RCC-AML in the same mass as AML only, Satoh et al.⁹⁾ as RCC only, while Schujman et al.¹⁰⁾

misdiagnosed coexisting RCC-AML in the same mass as transitional cell carcinoma in the upper calyx of the kidney. Even when RCC and AML were separated, correct preoperative diagnosis was made after the report of Takeyama et al.⁶⁾ in 8 of 14 cases (57%). At present, it seems that none of the diagnostic techniques listed above, nor a combination of some or all methods can resolve this problem, and only pathologists can correctly diagnose this kind of mixed tumor. Further advances with newer and sophisticated imaging techniques such as US, CT, and MRI may resolve this problem in the future.

Currently, a growing number of urologists are reporting excellent results with nephron-sparing surgery for patients with RCC. Licht and Novick³⁰⁾ summarized data on 241 patients in whom a mean disease-specific survival rate of 95% was obtained after an approximately three-year followup.

Although radical nephrectomy remains the accepted standard treatment for localized RCC with normal contralateral kidney, nephron-sparing surgery may be indicated in cases with small tumors.

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和文抄録

同一腎に腎細胞癌と腎血管筋脂肪腫を合併した 1 例

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症例は54歳女性。結節性硬化症の合併は認めない。腹部超音波検査にて、右腎の中部外側に高エコーを呈する腫瘤を、下極付近に前者よりもさらに高エコーを呈する腫瘤と指摘され当科受診。CT, MRI, 血管造影にて、右腎中部外側の腫瘤は腎細胞癌と、右腎下極付近の腫瘤は腎血管筋脂肪腫と術前診断された。1990年3月経腹膜的根治的右腎摘除術を施行、中部外側の腫瘍は腎細胞癌、alveolar type, common type, clear cell subtype, G1 と、また、下極付近の腫瘍は、腎血管筋脂肪腫と病理診断された。術後経過は良

好で、術後61カ月再発転移を認めない。われわれの経験した症例のように腎細胞癌と腎血管筋脂肪腫が同一腎内に別々の腫瘤を形成している場合は、Takeyama, Arima, Sagawa and Sonoda の報告以降、比較的正しく術前診断がなされている。しかし同一腎内の同一腫瘤内に腎細胞癌と腎血管筋脂肪腫が共存している場合には、正しい術前診断がなされたという報告は見あらず、今後に残された課題と思われる。

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